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SHORT REPORT

Spontaneous Regression of a Limb Arterio-Venous Malformation (AVM) in a Patient with Parkes-Weber Syndrome (PWS)

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Abstract Klippel Trenauney Syndrome (KTS) consists of the triad of venous and cutaneous capillary malformations, and tissue hypertrophy. The association with an Arterio-Venous Malformation (AVM) is known as Parkes-Weber Syndrome (PWS). We present a case of spontaneous regression of multiple small AVMs in the lower leg of a 19-year-old female with PWS. To the best of our knowledge this is the first documented case of spontaneous regression of an AVM in PWS in English literature.

Introduction

Parkes-Weber Syndrome was first described in 1907 by the physician Parkes Weber. It involves the triad of symptoms seen in Klippel Trenauney Syndrome — venous malformations, cutaneous capillary malformations and lymphatic malformations — in association with an AVM. This commonly occurs with limb hypertrophy secondary to soft tissue and bone overgrowth.[1] Recently the term haemolymphatic malformation has been used to describe such syndromes.[2]

The AVM in PWS is defined as a Congenital Vascular Malformation (CVM). The AVMs originate during embryogenesis, and they are not known to regress spontaneously.[2] Here we describe the spontaneous regression of an AVM in a patient with PWS.

Case History

The patient was born with hypertrophy of the left leg with associated port wine stain. At age two years an angiogram demonstrated abnormally dilated veins but no AVMs.

At age eight she had laser therapy for her port wine stain and had ligation of perforating veins for painful varicosities. A year later soft tissue overgrowth was limiting her function and an extensive debulking procedure was undertaken.

She subsequently developed various postural problems thought to be secondary to discrepancy in leg length due to osseous hypertrophy of the affected leg. At age 12 she had an L5/S1 spinal fusion for spondylolisthesis.

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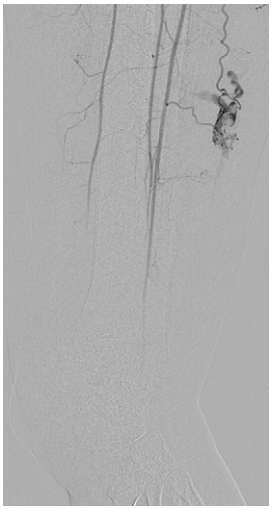

Figure 1 Percutaneous transfemoral angiogram demonstrating peroneal AVM.

At age 18 she was referred to our vascular unit because of pain in the left leg which was limiting her mobility. Examination revealed a grossly swollen left lower limb, dilated varicosities, and lipodermatosclerosis. MR angiography then demonstrated multiple abnormal connections between the arterial and venous systems, and a percutaneous transfemoral angiogram confirmed the presence of small AVMs arising from the proximal and distal peroneal arteries (Fig. 1).

As her symptoms deteriorated, the decision was made to treat the AVM and a repeat angiogram was undertaken two months later. Despite the use of pump injectors no AVMs were found and a normal arterial system was noted (Fig. 2). Venography demonstrated persistence of the venous malformation forming a complex meshwork of veins around the foot and ankle.

As her symptoms persisted, the venous malformation was treated with sclerotherapy. Repeat venogram and duplex scan two months following treatment confirmed obliteration of the venous malformation with an improvement in pain.

Discussion

Spontaneous regression of AVMs is rare. There are around 75 cases of spontaneous regression of cerebral and spinal AVMs in the world literature.[3] However, published reports generally relate to AVMs formed by a different aetiological process to that seen in PWS. Postulated reasons for AVM resolution include thrombosis secondary to contrast and trauma.[3,4]

In this case the spontaneous regression of the AVM could be because of a different pathophysiological origin to that of other AVMs seen in PWS, contrast angiography, or trauma (although there was no history). A false positive angiogram and MR angiogram are also possible, for example due to hyperperfusion secondary to infection, but this seems unlikely given the gap of several months between the investigations.

Figure 2 Angiogram using injector pump demonstrating resolution of AVM.

We report a case of spontaneous regression of an AVM in a limb in a confirmed case of PWS. To the best of our knowledge there is no other report in English literature. As has been reported by other authors, treatment of venous malformations helped to relieve symptoms in our patient.

Conflict of Interest/Funding

None.

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